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Autosomal dominant polycystic kidney disease [ADPKB] is one of the commonest genetic diseases. Apart from the involvement of the kidneys, several other organs, viz. the liver, the central nervous system, the pancreas, the spleen, the ovaries and the gut, amongst others, are also sometimes involved. This makes ADFKD more of a systemic rather than an isolated renal disorder. This becomes more so considering that the involvement of the other organs contribute significantly to the morbidity and mortality of ADPKD. This review looks at the pattern and prevalence of involvement of other organs, apart from the kidney in ADPKD.